

6.12 INTERSTITIAL LUNG DISEASE (ILD)

There are over 200 forms of interstitial lung disease (ILD). These conditions are characterised by increasing shortness of breath and widespread shadowing on chest x-ray. The most common are sarcoidosis and idiopathic pulmonary fibrosis (IPF). Approximately 55 new cases of IPF and 121 new cases of sarcoidosis are expected in Northern Ireland each year. ILDs with a known cause include asbestosis, bird fancier's lung and farmer's lung. ILD is also found in association with connective tissue diseases such as rheumatoid arthritis and scleroderma.

The investigation of a person suspected of having ILD should include a clinical assessment by a specialist respiratory physician. Specialised breathing tests are needed which should be carried out by trained respiratory physiologists. CT scanning is also usually required and the scans should be interpreted by radiologists with appropriate training in respiratory radiology.

Biopsies of the lung may be performed for diagnosis in some patients. These may be obtained through a fibre optic bronchoscope or by keyhole or open surgery through the chest wall. The results of biopsies should be interpreted by a histopathologist with an interest in respiratory pathology and in the light of discussion with the referring clinician.

The treatment of ILDs may include corticosteroids and cytotoxic drugs which can have serious side effects so that their use needs to be monitored with particular care. Further guidance is anticipated from NICE for the management of steroids. The inclusion of a standard on the treatment of ILD will be considered once this information becomes available.

Some types of ILD can progress and cause shortness of breath which does not respond to drug treatment. Specialist respiratory teams should be available to patients to help deal with the palliative care needs of these patients and those of their carers.

Overarching Standard 45:

Diagnostic Processes for Interstitial Lung Disease

All patients with ILD or suspected ILD should be under the care of a respiratory physician with appropriate clinical, physiological, radiological, pathological, surgical and laboratory support.

Rationale:

ILDs comprise a very wide spectrum of pathologies, presentations and outcomes. Patients with these conditions (or suspected of having these conditions) should be diagnosed and managed by staff with the appropriate training and skills. They should be under the containing care of a respiratory physician. There should be a designated regional lead in ILD available to provide tertiary support and to develop and coordinate services.

Lung biopsy samples should be obtained when the diagnosis remains uncertain after clinical and radiological assessment, unless there are patient contraindications or when the samples are unlikely to contribute to management.

Evidence:

British Thoracic Society Interstitial Lung Disease Guideline Group. Interstitial Lung Disease Guideline. Thorax, 2008;63 (Suppl 5): v1-v58.
doi:10.1136/thx.2008.101691 <http://www.brit-thoracic.org.uk/Portals/0/Clinical%20Information/DPLD/Guidelines/Thorax%20Sept%202008.pdf>

Responsibility for delivery / implementation

HSC Board
Public Health Agency
HSC Trusts (including respiratory physicians, clinical physiologists, radiologists, pathologists and thoracic surgeons)

Quality Dimension

1. All patients suspected of having ILD should be referred to a respiratory physician for assessment and management.
2. All patients with ILD should have access to a multidisciplinary team based in a regional centre with expertise in ILD.
3. All respiratory physicians should have on-site pulmonary function testing conforming to BTS/ARTP standards including spirometry, lung volumes and transfer factor and should also have ready access to cardiopulmonary exercise testing. Pulmonary function testing should be performed by a clinical physiologist with training in respiratory physiology.
4. Patients referred for high resolution CT scans of the chest should have the scans performed, reported and discussed with the referring physician within three weeks.
5. All respiratory physicians should have ready access to bronchoalveolar lavage (BAL), endo- and transbronchial lung biopsy and thoracic surgical procedures including video-assisted thoracoscopy (VATS) and open lung biopsy.
6. Patients who have biopsies for the investigation of ILD should have the results discussed by the referring clinician and the pathologist within two weeks of the procedure.

Performance Indicator	Data Source	Anticipated Performance Level	Date to be achieved by
Percentage of patients who have spirometry and gas transfer measurements at presentation	Trust ILD Database (to be developed)	Establish baseline Performance level to be determined once baseline established	March 2011
Percentage of patients who have had high resolution CT scans where these have been seen and discussed between the referring physician and the radiologist within three weeks	Trust ILD Database (to be developed)	Establish baseline Performance level to be determined once baseline established	March 2011